Almost a quarter of all birth defects — approximately one out of every 500 births — are the result of a congenital malformation of the kidneys and/or urinary tract. Research shows that diagnosing these congenital abnormalities in utero, and then following up with specialized long-term care shortly after birth, results in more children living longer, healthier lives.

A unique program in UCLA’s Division of Pediatric Nephrology offers specialized care for children with renal and/or urinary tract abnormalities beginning when an abnormality is detected in utero, continuing with treatment starting shortly after birth and including follow-up care as necessary throughout childhood.

Diagnosis in utero and carrying to full term

Renal and urinary tract abnormalities are often first discovered during a routine fetal ultrasound, which can detect anatomical abnormalities such as a solitary kidney or an extra ureter. Expectant mothers referred to UCLA’s pediatric nephrology program meet with a nephrologist to discuss kidney development in utero and strategies to prevent further damage to the baby’s kidney.

Teaching older children to be vigilant about their health

At some point, children with kidney conditions need to learn to be vigilant about their own health. “Usually between the ages of 8 and 10, parents will talk with their children,” says Patricia Weng, MD, assistant professor of pediatric nephrology. Helping these children adapt to living with their condition is a team effort involving the pediatric nephrologist, primary care physician, social worker and parents.

“It is important for these patients to feel like normal kids, but to also know that they have to be careful because, for example, they have only one kidney left, or they have another chronic condition,” Dr. Weng says. “As they become teens, they need to know to continue their treatment for optimal health and to modify their lifestyle to avoid getting dehydrated, to drink alcohol only in moderation if at all, not to do drugs and to be careful about taking medications such as NSAIDs, even for menstrual cramps.”
It is critically important to strive for a full-term delivery to allow the organs time to develop. Kidneys are not fully formed until the fetus is 36 weeks of age, and they continue developing throughout childhood. Expectant mothers are typically advised on optimal fluid intake and avoiding medications, including nonsteroidal anti-inflammatory drugs (NSAIDs), that reduce blood flow to the kidneys. After this session, the expectant mothers do not usually return to see the nephrologist until after they give birth.

**Monitoring and treatment of newborns**

While some kidney abnormalities resolve themselves before birth, others can have life-threatening consequences if not managed properly. The diagnosis should be confirmed soon after birth so appropriate care can be given.

Immediately after birth, diagnostic imaging studies are used to assess newborns’ kidney function. The first imaging test done is usually a renal ultrasound, which can assess the kidney anatomically and poses no radiation risk to the newborn. If it appears on ultrasound that a kidney is not functioning, a nuclear medicine scan will be conducted to check kidney function. If a ureter appears enlarged, a voiding cystourethrogram (VCUG) — a fluoroscopic exam of the bladder and urinary tract — is performed. VCUG can detect abnormalities in the flow of urine through the lower urinary tract, such as vesicoureteral (VU) reflux, in which urine passes upward from the bladder back into the kidneys via the ureter.

One of the most common congenital abnormalities of the kidney is dysplasia, which occurs when the internal structures of the baby's kidney do not develop normally. Network tubules that collect urine are not fully branched out and the urine becomes trapped in the kidney, causing fluid-filled cysts that replace normal tissue. Dysplasia usually affects a single kidney; with proper medical management, these children can grow normally and lead typical lives. These patients are monitored closely to control blood pressure and electrolytes. Babies with dysplasia of both kidneys who survive gestation will need dialysis and kidney transplantation very early in life.

Kidney agenesis, also known as solitary kidney, is another common abnormality. Studies show that children born with one kidney who are followed closely by a nephrologist experience fewer complications such as hypertension or proteinuria (excessive protein in the urine).